

ORSETT BRIEFING PAPERS FOR PSYCHOLOGISTS

NO.14 - Inter-sex Conditions

INTRODUCTION

"Gender identity" is how the individual feels about themselves (ie: male, female; masculine, feminine, androgynous). "Biological sex" is the sexual identity of the individual based on their biological make-up (ie: biological male, biological female, inter-sex, hermaphrodite).

Biological sex is determined at a number of levels ¹ (table 1).

	<u>MALE</u>	<u>FEMALE</u>
1. Chromosomes	XY	XX
2. Reproductive organs	testes	ovaries
3. Dominant hormones	testosterone	oestrogen/ progesterone
4. Internal organs that link reproductive organs and external genitalia	Wolffian duct	Mullerian duct
5. External genitalia	penis	vagina/clitoris

Table 1 - Levels of determining biological sex.

Usually these levels work together, but in exceptional cases, they may be different due to sex chromosome or hormonal abnormalities.

Table 2 gives a classification of inter-sexual disorders (some of them exceptionally rare).

¹ Hutt, C (1978) Sex role differentiation in social development. In McGurk, H (ed) Issues in Childhood Social Development London: Methuen.

<u>CONDITION</u>	<u>DESCRIPTION</u>
congenital adrenal hyperplasia (adrenogenital syndrome)	XX (genetic female) with excess androgens (hormones) in womb; male appearance of genitals; eg: enlarged clitoris and fused labia
Turner's syndrome	X0 (no second X chromosome); generally female appearance of genitals
Klinefelter's syndrome	XXY (male with extra X chromosome); male with small penis and rudimentary testes
"Supermales"	XYY (extra Y chromosome); eg: taller
Androgen insensitivity syndrome (AIS) (testicular feminizing syndrome)	XY (genetic male); external genitalia appear female; can have breasts in extreme cases
Enzymatic defects in XY genotype	Interruption in testosterone in womb; ambiguous or female looking genitals ²
Hermaphroditism	Both testes and ovaries; 60 cases in past 100 years in Europe and North America ³
Pseudohermaphroditism	XX with masculine-looking genitals; XY with rudimentary testes
"Superfemales"	XXX ("super-X syndrome"); XXXX; XXXXX; XXXY ⁴
5-alpha-reductase deficiency	Problem with testosterone similar to AIS ⁵ ; assumed to be female at birth but penis develops at puberty; called "kwolu-aatmwol" (New Guinea) meaning "female thing-transforming-into-male thing" ⁶ ; "guevodoces" (Dominican Republic) meaning "balls at twelve" ⁷

² Sadock, B.J & Sadock, V.A (2003) Kaplan and Sadock's Synopsis of Psychiatry (9th ed) Philadelphia: Lippincott Williams & Wilkins.

³ Money, J (1986) Venuses Penuses: Sexology, Sexosophy and Exigency Theory Buffalo, NJ: Prothemeus.

⁴ Caroline Cossey - male to female trans-sexual - speaking on "Open to Question" 1992; BBC Television).

⁵ Mealy, L (2000) Sex Differences: Developmental and Evolutionary Strategies San Diego: Academic Press.

⁶ Stainton Rogers, W & Stainton Rogers, R (2001) The Psychology of gender and Sexuality Buckingham: Open University Press.

⁷ Imperato-McGinley, J et al (1974) Steroid-5-alpha-reductase deficiency in men: An inherited form of male pseudohermaphroditism Science 186, 1213-1215.

Table 2 - Different types of intersexual disorders.

The situation of inter-sex individuals or those with ambiguous genitalia is that they tend to have a "somewhat vagina like opening" and "a phallic structure of a size and shape that could be described as either an enlarged clitoris.. or a micropenis" ⁸.

Surgery is performed on newborns based on the decision of which sex to raise the child. If the penis is less than one inch (2.5cm) at birth, then the tendency is to perform surgery to "make" female, and if the clitoris is larger than one quarter of an inch (0.9cm), the decision will be to "make" male ⁹. About 2000 babies in the US each year have some kind of surgery for ambiguous genitalia ¹⁰.

Kessler ¹¹ questioned the assumptions about "ambiguous genitals": "They (obstetricians) seem to know ambiguity when they see it.. the phrase 'ambiguous genitals' is used freely with no apparent need to define what 'ambiguous' means in this context". Furthermore, she said: "Ambiguity is at least partially determined by factors such as who is looking, why they are looking, and how hard they are looking.. How hard one 'looks' at genitals and what one 'sees' is not constrained by the optic nerve but by ideology".

SEX CHROMOSOME ABNORMALITIES

Sex chromosome abnormalities lead to a number of conditions, of which the main ones are:

- Turner Syndrome

Develop as female, but lack normal ovaries, which can mean no puberty. Physically short with unusually broad chest and loose skin around the neck. Frequency of live births: 1 in 4000 ¹².

- Klinefelter Syndrome

Develop as male, but with some degree of learning disability. Often extreme tallness, and have breast development (gynecomastia). Frequency of live births: 1

⁸ Colapinto, J (2000) As Nature Made Him. New York: HarperCollins p216.

⁹ Baird, V (2001) The No-Nonsense Guide to Sexual Diversity. London: New Internationalist.

¹⁰ op cit Sadock & Sadock.

¹¹ Kessler, S.J (1998) Lessons from the Intersexual. New Brunswick, NJ: Rutgers University Press.

¹² LeVay, S & Valente, S.M (2006) Human Sexuality (2nd ed). Sunderland, MA: Sinauer Associates.

in 1000 ¹³.

- XYY Syndrome

Develop as male, but with genital anomalies. Learning disability, and increased rate of criminal convictions ¹⁴. Frequency of live births: 1 in 1500 ¹⁵.

- Triple X Syndrome

Develop as female, but very little difference apart from occasional cognitive deficits. Frequency of live births: 1 in 2000 ¹⁶.

HORMONAL ABNORMALITIES

Hormonal abnormalities in the womb lead to four main inter-sex conditions:

- Androgen Insensitivity Syndrome (or Testicular Feminization)

Genetic mutation ¹⁷ makes womb insensitive to androgens (eg: testosterone) (complete AIS), and partial AIS has limited insensitivity. The chromosome pattern is XY (male), but lack of testosterone limits the development of male genitals. The physical appearance is female, but no menstruation at puberty. Individuals with CAIS have female external genitalia and normal testes. Frequency in liveborn males in Danish study was 1 in 20 400 ¹⁸.

A Spanish female athlete, Maria Patino, at the 1992 Olympic Games was disqualified for having a XY chromosome pattern, but she had no idea herself and had been raised female ¹⁹.

MALE (XY) → FEMALE GENITALIA APPEARANCE

¹³ *ibid.*

¹⁴ Gotz, M.J; Johnstone, E.C & Ratcliffe, S.G (1999) Criminal and antisocial behaviour in unselected men with sex chromosome abnormalities Psychological Medicine 29, 953-962.

¹⁵ *op cit* Levay and Valente.

¹⁶ *ibid.*

¹⁷ A "loss-of-function mutation" in the androgen receptor on the X chromosome (Wilson 2006a).

¹⁸ Wilson, B.E (2006a) Androgen Insensitivity Syndrome (www.emedicine.com/ped/TOPI2222.HTM; accessed last on 30/04/08).

¹⁹ Bown, W (1999) Sex-test confusion could create havoc at Olympics New Scientist 18/1, p14.

- Gonadal Intersexuality

This is "true" hermaphroditism. Individuals possess both testicular and ovarian tissue usually seen as one ovary and one testis. Usually XX chromosomes (female), but something has happened in womb to produce testis (eg: SRY gene translocated to X chromosome).

Another possibility is that two sperm (one X and one Y) have penetrated a single ovum, but only one of them has fertilised the egg and the other has affected the development in some way.

A famous case known as "Mr.Blackwell" was an individual who had a vaginal opening and a penis, an active ovary on one side of the body and an active testicle on the other ²⁰.

- Congenital Adrenal Hyperplasia (or adrenogenital syndrome)

Individuals with XX chromosomes (female) who through a genetic defect receive too much androgens in the womb and after birth leading to masculinity and ambiguous genitalia (eg: enlarged clitoris).

There is a "classic" version caused by 21-hydroxylase deficiency, and a milder, "non-classic" version. 21-hydroxylase is an enzyme which if lacking causes the body to overproduce androgens in the womb ²¹.

It is a recessive gene condition which can remain in certain genetic populations (eg: Ashkenazic Jews from Central Europe - 1 in 27 children have mild version ²²). The frequency of live births in the general population for the "classic type" is 1 in 16 000 ²³.

The condition can also affect XY fetuses, but is only noticeable after birth as early puberty.

FEMALE (XX) → MALE GENITALIA APPEARANCE

²⁰ Goldwyn, E (1979) The fight to be male Listener May 24, 709-712.

²¹ Speiser, P.W (2005) New Developments in the Treatment and Diagnosis of Congenital Adrenal Hyperplasia (www.medhelp.org/nadf/nadf5.htm; last accessed 30/09/07).

²² New, M.I & Wilson, R.C (1999) Steroid disorder in childhood: Congenital adrenal hyperplasia and apparent mineralocorticoid excess Proceedings of the National Academy of Sciences of the USA 96, 12790-12797.

²³ Carlson, A.D et al (1999) Congenital adrenal hyperplasia: Updates on prenatal diagnosis and treatment Journal of Steroid Biochemistry and Molecular Biology 69, 19-29.

- 5-Alpha-Reductase Type 2 Deficiency (5-ARD)

A recessive gene leads to problems with an enzyme (5-alpha-reductase) which interacts with testosterone. It is very rare except for clusters in limited genetic populations (eg: Salinas in Dominican Republic ²⁴; specific populations in New Guinea and Turkey ²⁵).

The sufferers are male (XY), but the external genitalia appears female (through lack of testosterone in this aspect of development). The increased testosterone at puberty leads to full development of male genitalia.

MALE (XY) → AMBIGUOUS GENITALIA AT BIRTH → PUBERTY MAKES CLEAR MALE

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<http://www.emedicine.com>

²⁴ op cit Imperato-McGinley et al.

²⁵ Wilson, B.E (2006b) 5-Alpha-Reductase Deficiency (www.emedicine.com/ped/topic1980.htm; last accessed 30/04/08).

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